PSEUDOPELADE OF BROCQ: ITS RELATIONSHIP TO SOME FORMS OF CICATRICIAL ALOPECIAS AND TO LICHEN PLANUS

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CONCEPT AND DELIMITATION OF THE PSEUDOPELADE SYNDROME

This disease was first described by Brocq, who reported the first case known in a letter addressed to the "Journal of Cutaneous and Veneral Diseases" in 1885, but it was not until 1907 that his excellent description of the clinical characteristics of this disease, appeared in his "Traité de Dermatologie Pratique" (page 648). These were later confirmed by Pautrier, Sabouraud (1), Photinos (2) and by many other authors.

In his descriptions, Brocq insistently repeats the complete absence of clinical inflammatory phenomena, a characteristic that is useful to differentiate this process quite definitely from some varieties of inflammatory folliculitis described at about the same time by Quinquaud under the name of "Folliculite décalvante" and by Lailler under the name of "Acné décalvante". In reality the different stages of transition between both diseases do not permit such a precise differentiation, and as Degos, Rabut, Duperrat and Leclerq (3) suggest in a recent excellent publication, many authors confuse both syndromes considering the alopecia maculosa, which is the pseudopelade, as the final stage of some forms of pustular folliculitis.

In a survey of their material in 1947, Miescher and Lenggenhager (4) conclude that a certain amount of papillary reddening is a clinical symptom commonly found in the initial stages of pseudopelade of Brocq, and that frequently a certain degree of desquamation can be observed.

Brocq, in 1888 described under the name of "Sycosis Lupoide" a disease pertaining to the group of folliculitis decalvans, which was characterized by a chronic follicular inflammation which produced—after some time—a cicatricial atrophy in the form of smooth or shiny patches. In 1899 Unna described the same process with the name of "Ulerythema sycosiforme". This disease has a predilection to localize in the beard region, but may also appear on the scalp, with special predisposition to localize in the marginal line of the hair. Hoffmann (5) in an excellent publication describes in detail the clinical characteristics of this localization in the scalp, naming it "folliculitis sycosiforme atrophicans capillitti". Galewsky (6) makes a clear distinction between these two processes which from our point of view have no other real difference except that the sycosis lupoide of the scalp localizes electively in the marginal zone of the hair; but otherwise they have identical clinical characteristics.

There are, lastly, a series of atrophic alopecic affections of the scalp that simulate pseudopelade so much, that often they have been confused with them, es-

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pecially the atrophic cicatricial alopecia consecutive to the scalp localization lupus erythematosus and scleroderma. This justifies the concept of Degos and co-workers, who suggested the term "pseudopeladic state" instead of considering pseudopelade as an autonomous disease.

Finally in certain cases of pseudopelade the follicular funnel of the hairs that marginate the cicatricial alopecic patches show a slight degree of follicular hyperkeratosis, and may show in some cases, horny cones deeply rooted in the follicular funnels in place of the hairs that disappeared, as described by Galloway, Whitfield, Fox, Jackson, who gathered the facts published in the report by Brocq, Lenglet, and Ayricnac in 1905.

These follicular horny formations are very apparent in the so called Graham Little syndrome, which we shall discuss further on.

OUR CONCEPT OF LICHEN PLANUS

There has been some abuse, we believe, on the part of many authors regarding the exact concept of what is lichen planus. We believe it is necessary to state, adhering to the concept of our Professor Pautrier, that we understand by lichen planus or lichen ruber planus, the disease described by Erasumus Wilson in 1867 (7) whose name remained attached to this disease. It was precisely in England, where the clear concept of this disease was established, that in 1905, Radcliffe Crocker and Adamson (8) gave the name of lichen spinulosus to a trivial lesion of the pilosebaceous follicle, a process that can be found in a great variety of different diseases (Lichen scrofulosorum, peripilar syphilids, trichophytids, pityriasis rubra pilararis, certain deficiency states, certain intoxications, etc.).

Darier proposes the grouping together of all these cases characterized by a pure cutaneous reaction, under the name of "espinulosism." For the sake of brevity, we refer our readers who wish more extensive information about these points, to the excellent publication of Pautrier in "Nouvelle Pratique Dermatologique" (Volume VII, 1936) and to the "Précis de Dermatologie" of Darier.

THE GRAHAM LITTLE SYNDROME AND THE RELATIONS OF PSEUDOPELADE WITH LICHEN PLANUS

Graham Little described for the first time in 1915 (9) a syndrome characterized by cicatricial alopecia in patches over the scalp, together with a non-cicatricial alopecia of the axillae and pubic region, associated with an eruption of lichen spinulosus along the trunk. In this first report he did not consider the dermal lesions of the trunk as a true lichen ruber and only later (10, 11) admitted the thesis of Pringle grouping together the "lichen spinulosus" and lichen planus, and accepting the author's denomination of "lichen planopilaris".

The discussions which have arisen due to the confusion between what we believe are so clearly different disease entities, are very numerous and in the literature we find in opposition to Van Der Meiren's (12) well defended criteria disregarding the relation between "acné corneum", "keratosis pilaris" and Wilson's lichen, the opposing view recently defended vigorously by Spier and Keiling (13), in a well documented report with a complete bibliographical review, but

based upon a single personal observation where the dermal lesions were not at all similar to those found in "lichen ruber". Pierini and Borda (14) in 1949 reported seven clinical observations in which pseudopelade coincided with a typical cutaneous eruption and affirm that he "maintains the lichen nature of the dermatological picture known as pseudopelade of Brocq." They discuss the possible relation between spinulosism and lichen planus, accepting Adamson's point of view which considers that "the lichen spinulosus of adults is always followed by lichen planus". This would mean, according to Freund, that all cases of spinulosism in the adult are nothing more than a masked form of lichen planus, due to this peculiar hyperkeratotic reaction.

Following Pierini's publication, two different problems arose: 1) Is pseudopelade an authentic manifestation of lichen planus, as the described histologic characteristics and the coincidence of all his cases with a typical Wilson's lichen had proven? 2) Are all the cutaneous processes characterized by spinulosism true lichen planus? Pierini, abandoned this viewpoint, maybe without noticing it, when he praised Mottá's publication, in which the author classifies all the spinulous processes under the general heading of "folliculosis". If not, how can we admit that pityriasis rubra pilaris of Devergie, the follicular scurvy-like and the follicular syphilids, to cite only three examples, are similar to lichen planus?

With this confusion in the literature, it is very hard to determine in what proportion of cases pseudopelade and lichen ruber planus coincide. The facts stated by Barber, Beatty, Dore, Ormsby, Senear, MacCafferty, etc., cannot be taken into account, because of the habitual confusion that prevails among English speaking dermatologists, regarding lichen ruber and lichen spinulosus.

Degos and co-workers, who like us and the majority of French authors, make a clear cut differentiation between lichen planus and lichen spinulosus in a series of 100 cases of pseudopelade, find in six cases the coexistence of cutaneous lichen planus. On the other hand they notice in eleven cases the coexistence of horny cones, in five the association with pilar keratosis of the face and extremities; likewise the authors segregate quite clearly the Graham Little syndrome from the pseudopelade of Brocq.

PERSONAL EXPERIENCE

Pseudopelade

Gathering cases from our hospital service and private practice seen in the last years, we have been able to group 34 cases of pseudopelade, whose clinical characteristics correspond exactly to the classical description: the presence of alopecic patches of variable size, initially of small size, but by coalescence in cases of long duration they finally constitute patches as large as the palm of the hand, localized particularly in the vertex and upper part of the occipital and temporal regions. In all our cases, the alopecic surface was glistening white. In the small patches there was a slight depression and only in one half of the cases were we able to find a slight follicular hyperkeratosis that surrounded the hair, making the "ostium" appear slightly prominent and evidently darkened. Only in three cases did we find in some patches a slight furfuraceous desquamation and

in five cases of less than one year's duration we have found a slight reddening around some follicles.

We have not included in this group any cases of atrophic alopecia with marked folliculitis or any other cases with a previous dermatologic process of the scalp responsible for the alopecia resembling pseudopelade.

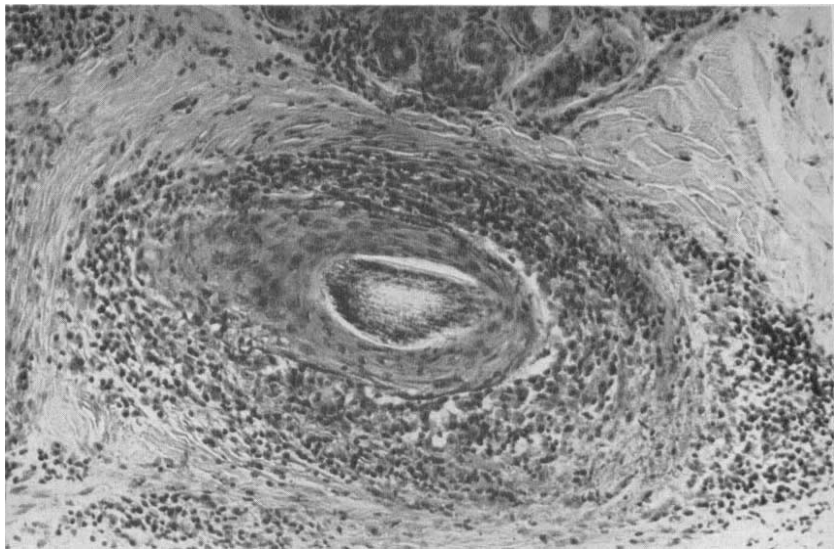

FIG. 1. Photomicrograph—perifollicular lesion of a typical case of pseudopelade. Lymphocytic perifollicular infiltration and slight edema.

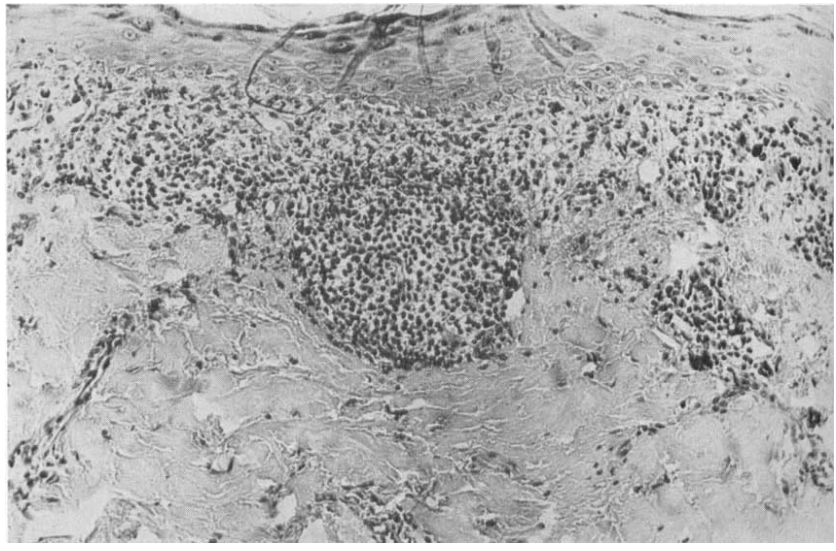

FIG. 2. Photomicrograph of another non-follicular lichen planus papule of a patient with typical pseudopelade without concomitant lichen planus. Scalp.

In our statistics pseudopelade constituted 0.65 % of all the patients seen in our Service during the same length of time. There was a marked affinity for females: 23 patients were women and only 11 men. However, much importance cannot be attached to this fact, because our clinical material consists of approximately 60 % women.

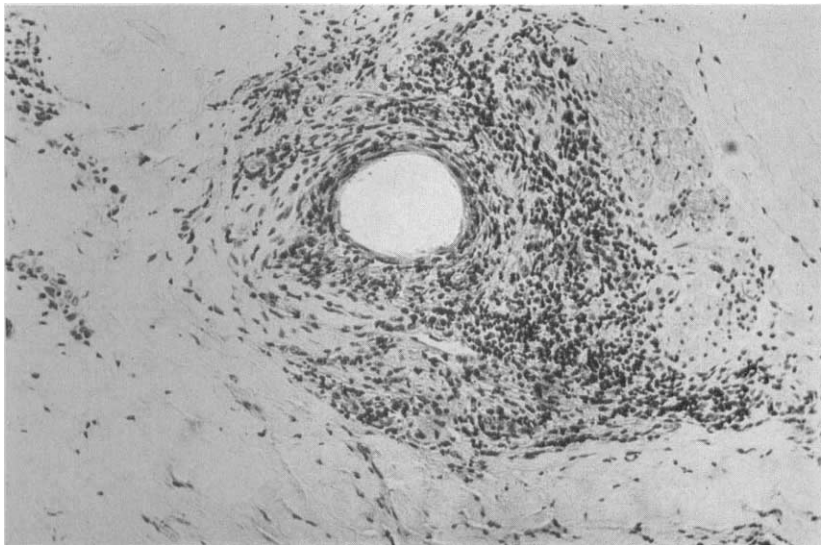

FIG. 3. Lymphocytic perifollicular infiltration around a pilosebaceous follicle undergoing atrophy, from the scalp of the same patient of previous microphotographs.

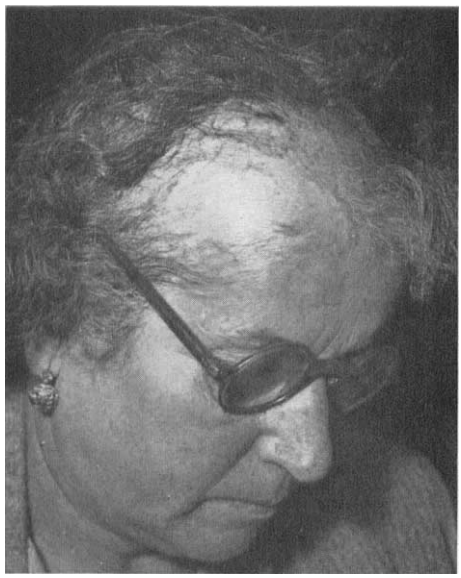

FIG. 4. Graham Little syndrome. Scalp lesions

The age of our patients varied from 18 to 54 years and the time of evolution of the disease when first reported was from seven months to fourteen years.

The course of all our cases has been irreversible, although it is possible that some forms of local treatment if regularly used, may have retarded the progressive extension of the lesion. As our co-workers Lopez Martinez and Cardenas (15) reported in 1948, we have never been able to observe phases of more rapid extension of the process separated by periods without change.

The detailed observation of the rest of the skin has permitted us to observe in two cases the simultaneous existence of a typical cutaneous lichen planus, in two cases lichen planus of the mucosa of the oral cavity and in one case a pigmentary alteration of the trunk that clinically could be diagnosed as "incontinentia pigmenti" although because of the disappearance of the patient we were not able to perform a histopathologic examination.

In no other case were we able to find any other concomitant cutaneous process nor any systemic disturbance that may have had any relation to this disease.

Histological study. In the previously mentioned report of Lopez Martinez and Cardenas that compiled our experience until 1948, the pathological characteristics were described in agreement with the majority of the authors, and coinciding with the excellent description of Civatte and Photinos, which we believe is not necessary to repeat. In the photomicrographs illustrating that work the essential characteristic histological picture can be seen.

In fig. 1 we see the characteristic dense lymphocytic infiltration surrounding slightly the superficial portion of a pilosebaceous follicle, showing a certain degree of edema; in the same picture we can observe a less marked infiltration around some blood vessels and around a sudoriparous gland.

In a biopsy performed on the scalp of a 30 year old man with a typical pseudo-

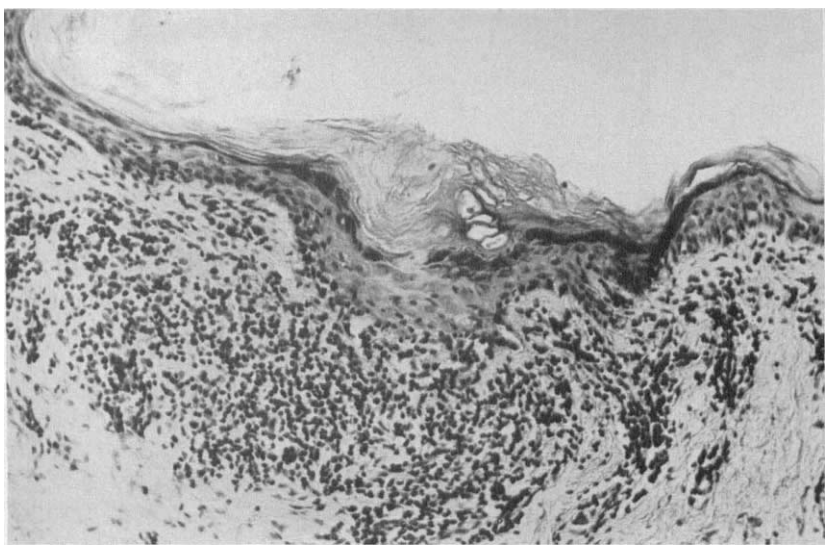

FIG. 5. Lichenoid papule of a reticular pigmented poikiloderma lesion from the neck of the previous case.

pelade that had begun three years before, and which clinically showed no association with lichen planus of the scalp, we were surprised to find lesions that were exactly similar to the papules of lichen planus. In the photomicrograph of this patient (fig. 2) we find find the following lesions: slight hyperkeratosis, hypergranulosis, disappearance of the normal arrangement of the basal cells, and a

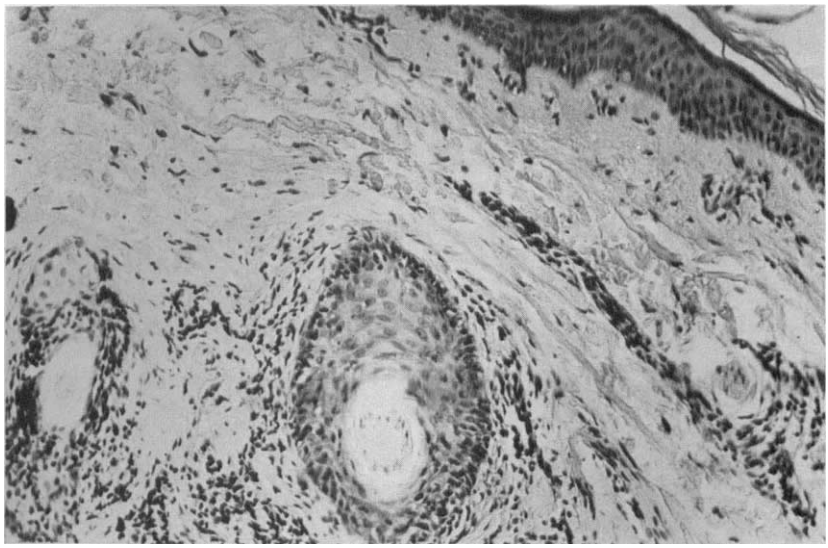

FIG. 6. Photomicrograph of the scalp of the same patient. Perifollicular lymphocytic infiltration, similar to pseudopelade.

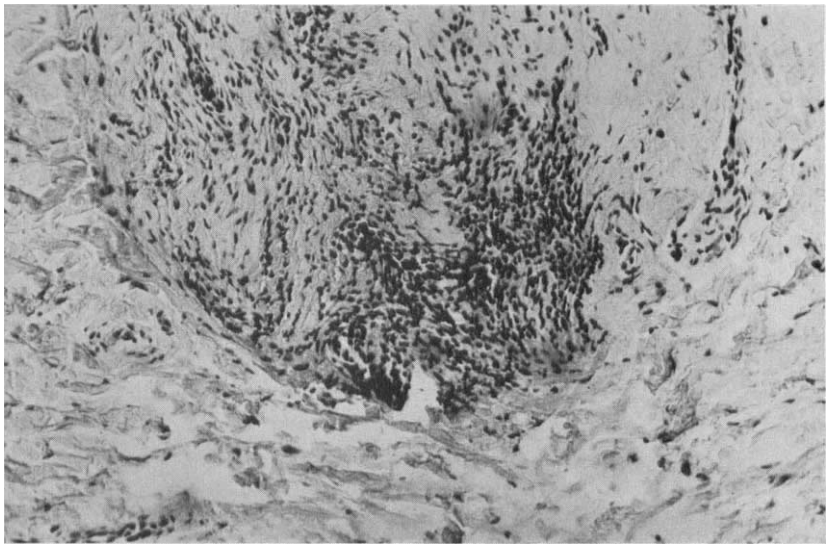

FIG. 7. The same patient with Graham Little syndrome. Follicular atrophy in scalp biopsy, showing a fibrous transformation of the follicle, similar to that seen in pseudopelade.

dense infiltration with lymphocytes with some melanophore cells in contact with the epidermis and a marked edema at the dermo-epidermal junction.

In this same patient we found a perifollicular infiltration, probably greater than in other cases, localized around some follicles undergoing a definite atrophic regression as may be evidenced in fig. 3.

Graham Little syndrome

We want to refer only to the following observation: a woman 53 years old came for consultation stating that three months ago she suffered an alimentary intoxication of unknown origin and since then she noticed a spinolous eruption with intense pruritus localized on the trunk, especially about the shoulders and the upper part of the extremities. There were also some pigmented patches on the lateral parts of the neck that extended up to the lower part of the face.

The dermal lesions consisted of typical spinulosism; filiform, hard, horny prolongations, appearing over the normal skin which showed no change in color and no papular infiltration at its base.

FIG. 8. Neck of a patient with Graham Little syndrome consecutive to an arsenical lichen, after salvarsan. Note the existence of spinolous lesions.

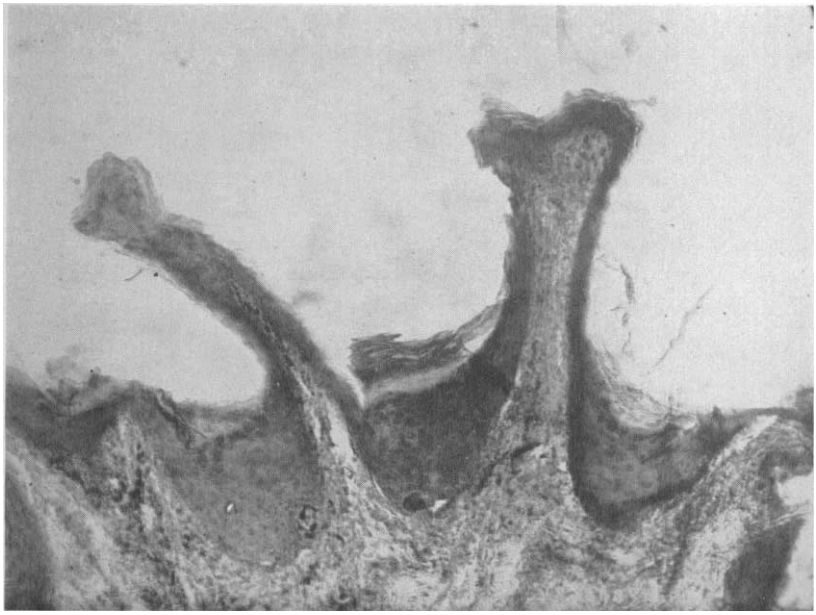

FIG. 9. Histological structure of the lesions of the previous figure. The spinolous process is due to a "papillomatosis" more than to the existence to follicular horny plugs.

In the scalp there were alopecic patches of considerable size that occupied the fronto-parietal and occipital regions. The greater part of the follicular orifices that had lost their hair, were filled up by a dark horny plug. (fig. 4).

On the lateral aspects of the neck, there was a dark reticular pigmentation that could be identified as a typical reticulated pigmented poikiloderma of Civatte.

In the axilla and pubic region there was a diffuse alopecia without horny cones.

The patient was subjected to a treatment with high doses of vitamin A. After four months she returned to us for observation and while all her lesions located on the trunk and lateral aspects of the neck were completely cured there was persistence in the scalp of cicatricial alopecic patches bordered by a diffuse alopecia with the presence of horny cones that had otherwise disappeared from the center of the patches which showed a smooth and glistening white surface. (fig. 4).

The histopathologic study of a biopsy taken from the lateral aspect of the neck showed the existence of follicular hyperkeratosis partially detached corresponding to one of the spinolous hyperkeratoses, and below, were, the small typical lichenoid papules described as characteristic of the reticulad pigmented poikiloderma described by Civatte and Olga Eliascheff (fig. 5).

In the scalp we can see a discrete lymphoid infiltration around the follicles some of which are undergoing degeneration and in their place a fibrous structure is being organized around which there is a marked edema of the fibrous tissue (fig. 7). The horny cones, that envelop the remains of degenerated hairs, distend considerably the follicular orifices.

DISCUSSION

The concept of pseudopelade as an autonomous disease

Undoubtedly the clinical entity described by Brocq under the name pseudopelade and by Drew under the more proper name of "alopecia parvimaculata",

has an unquestionable clinical basis. Authors with the experience of Brocq, Photinos and Pautrier, do not even mention the possible relation of this process to other diseases, and they distinguish it completely from the depilating varieties of folliculitis, particularly from the "acné decalvante" of Quinquand.

Recently, the important publication of Degos and co-workers discusses 100 cases; in 39 the disease appeared in its classical form, not associated with any other disease of the skin or scalp. Of the 61 remaining cases, 39 coincided with other lesions of the scalp, some of which can be considered only as a fortuitous co-appearance of two different processes; the same thing can be said about the remaining 22 cases which showed coincident or healed lesions, such as hydradenomas of the eyelids, lupus erythematosus, scleroderma, Pick-Herxheimer disease, etc.

In our material, 28 cases presented a pure pseudopelade, clinical and histologically characteristic, without any preceding or concomitant dermal disease.

It is evident that the most difficult differentiation to establish is between pseudopelade and inflammatory folliculitis, especially with the clinical pictures described by Quinquand and Lailler. As Degos points out very well, there is no absolute opposition between these processes and the three authors that described these diseases for the first time (Brocq, Quinquand and Lailler) agreed fundamentally in that the first, namely pseudopelade was a milder form of these depilating forms of folliculitis and the other two, as the final stage of a clinically more apparent form of chronic folliculitis.

In the last survey published by Sabouraud, the author insists that sometimes there may be a slight almost unnoticeable peripilar reddening in the recently formed patches of pseudopelade, this being a clinical sign of the existing folliculitis. This same has been underlined by Miescher and Lenggenhager.

The majority of authors agree, with but slight differences, about the histologic description of the lesions. Because of lack of space we shall not enter into detailed description of the pathology, but it is quite evident that the perifollicular inflammation, typical of pseudopelade, and which may vary in degree, has been interpreted by some authors (Pierini and Borda) has proof that it was really a follicular papule of lichen planus. The histologic differences between both processes may really be very slight especially in different sections and perhaps they have been stressed inordinately as an argument to defend an obstinate thesis.

Pseudopelade and lichen planus

We mentioned previously, that Pierini and Borda in 1949 reported seven cases in which pseudopelade appeared concomitantly with lichen planus, stating that "the dermatologic process known as pseudopelade of Brocq has a lichenoid nature". In a publication by the same authors two years later (16), they report 11 new cases, the majority of them, according to our concept, could not withstand strict critique. Especially in the photomicrographs 1 and 2 illustrating their report, we can see the typical and well known histologic lesions of pseudopelade of Brocq, but we believe that they cannot be used as a definite argument to affirm the lichenoid nature of the processes mentioned.

We have already stated how difficult it is to study the literature about this specific disease, because there is much confusion about "spinulosism" and "lichen ruber planus". Above we clarified our concept about the latter disease, in accordance with the concept supported by the French school of Dermatology and by a great number of German dermatologists.

That is why we shall mention the observation of Degos and coworkers in which they find 6 cases (in a group of 100) in which the disease appears in association with a typical lichen planus. In our personal experience, we found this association four times in 34 cases.

By chance we had the opportunity to observe a very interesting finding: a 30 year old man, who never presented lichen planus lesions, came to our service with a typical pseudopelade that had appeared three years before. In a histologic study of a scalp biopsy we found some lesions that were not follicular and were completely identical with the papules of lichen ruber planus. (Fig. 2).

From these facts we are inclined to deduce that the association of pseudopelade and lichen planus is too frequent to be considered as purely casual; but it would be an exaggeration to state that all the cases of pseudopelade are related to lichen planus of the scalp.

Graham Little syndrome and Pseudopelade

We believe that the clinical existence of this syndrome is unquestionable. We therefore want to report in this paper a personal observation of a typical case.

In accordance with Degos' observation, the initial clinical picture found in the scalp can not be confused with pseudopelade, but when the horny cones disappear and only the flat cicatricial alopecia remains, the similarity with old patches of pseudopelade is striking.

In other observations of the Graham Little syndrome, for example, in a case of arsenical lichen, after salvarsan, we were able to see the same spinolous lesions on the trunk, as we can see in fig. 8, but we have found completely different histologic lesions in this type of case. By comparison we can see how these spinolous lesion in the first case consist of a follicular horny plug and in the second case of protruding papillary prolongations with hyperkeratosis at the apex (fig. 9).

It is quite evident that the histologic lesions seen in fig. 5, can be interpreted as lichen planus papules, an argument in favor of the lichenoid etiology mentioned in recent publications by Graham Little. But it is also evident that these lesions coincide entirely with the small "lichenoide" papule described by Eliascheff, who found them in reticular poikilodermas and considered them as pathognomonic of this process which has never been confused with lichen planus.

In 1953 Spier and Keilig arrive at the conclusion that pseudopelade, for which they propose the name of "lichen ruber follicularis decalvans" is nothing but an abortive form of the Graham Little syndrome localized exclusively in the scalp. They base their assertion on a single personal observation, where the dermal lesions are very difficult to evaluate from the illustrations in their report, and which seem morphologically similar to "lichen scrophulosorum". They called them, "lichen ruber follicularis lanuginis".

The authors, commenting on a report of Degos, Rabut, Duperrat and Leclerq, sum up in the same group the cases associated with lichen planus and with follicular keratosis on the body or in the patches of pseudopelade, thus stressing the coincidence of pseudopelade and lichen planus.

In reality, these authors, in a later report of 100 cases of pseudopelade, besides the 7 cases associated with lichen planus which we mentioned before, report that eleven cases showed horny cones at the level or at the margin of some alopecic patches, and five cases showed keratosis pilaris of the face and extremities and in one case a Graham Little syndrome. That is to say, that the authors establish a clear difference between the Graham Little syndrome and the simple presence of horny cones or keratosis pilaris.

In conclusion, we want to mention that if we accept the concept of Graham Little, Pringle, Spier and Keiling and all the authors who, like Piérini and Borda, consider the Graham Little syndrome, the so called lichen pilaris planus as identical with the presence of horny cones in the pseudopelade patches, a great confusion will undoubtedly be produced. By a careful consideration of the observations mentioned by these authors we cannot establish a precise division between typical lichen ruber planus, a disease which all dermatologists would agree in diagnosing, and the poorly defined and heterogenous group of keratosis pilaris or folliculosis, following Motta's classification.

In the pseudopelade patches, we can find with relative frequency (11% in Degos' statistics) some follicular horny plugs, especially at the margin of the alopecic patches. More often we can find a slight reddish tint around the hairs at the border of the alopecic patches, an almost imperceptible elevation of the ostium follicularis, which histologically corresponds to a slight follicular hyperkeratosis, a mild reaction that accompanies practically all cicatricial alopecic processes of the scalp. A great number of people may show, in some region of the body, more or less pronounced lesions of keratosis pilaris to which we attach no importance, and even though they may be concomitantly affected by pseudopelade, no dermatologist would consider them as cases of lichen planus. In the majority of papers from authors who defend the exclusive lichen nature of pseudopelade we can observe over-emphasis in order to demonstrate their point, confusing the disease with abortive forms of lichen planus.

CONCLUSIONS

In summary, we can conclude that pseudopelade is the final and irreversible stage of a follicular process of the scalp, which follows a chronic course, sometimes being clinically evident and at other times made apparent only on histologic study.

A number of diseases of the scalp, such as chronic suppurative folliculitis, lupus erythematosus, scleroderma, some forms of congenital alopecias, the Graham Little syndrome, etc., may produce in the final stages some forms of cicatricial alopecia clinically identical with pseudopelade.

The association of pseudopelade with lichen planus is not infrequent and at times we can find a typical lichenoid histology in the patches of pseudopelade.

For all these reasons, we should accept Degos' proposal to consider these processes as "pseudopeladic states", which to us appears to be a very suitable term.

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